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Generalized lichen nitidus in a 6-year-old girl with Down syndrome

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ABSTRACT

Lichen nitidus is a generally asymptomatic disease identified by shiny, flesh-colored papules that are often limited to the trunk, upper limbs, and genitalia. Here, we describe a generalized presentation of lichen nitidus in a 6-year-old girl with Down syndrome. Lichen nitidus shares a known association with Down syndrome, and several case studies document patients with Down syndrome developing generalized lichen nitidus. The extensive nature of our patient's lichen nitidus, as well as the uncommon distribution, particularly on the face, adds to the currently limited primary literature on this subject.

KEYWORDS Down syndrome; generalized lichenoid eruption; lichen nitidus

Lichen nitidus is an uncommon inflammatory skin condition characterized by typically asymptomatic flesh-colored papules 1 to 2 mm in diameter. Lesions are usually localized to the trunk, upper limbs, and genitalia. Although lesions due to lichen nitidus are usually asymptomatic, some patients report pruritus, requiring treatment.[1] Lichen nitidus tends to affect children and adolescents more than adults and does not appear to demonstrate a preference for any race or gender. Though the exact pathophysiology of lichen nitidus is unknown, it has been known to co-occur with lichen planus, erythema nodosum, segmental vitiligo, and lichen spinulosus.[2] Several published case reports identify cases of generalized lichen nitidus in children with Down syndrome.[3–9] Here, we document another case of generalized lichen nitidus in a child with Down syndrome and synthesize findings from relevant cases. We also share photographed improvement of lesions at 3-month follow-up, something not previously documented in children concurrently affected by lichen nitidus and Down syndrome.

CASE REPORT

A 6-year-old black girl with Down syndrome presented to the dermatology clinic upon referral by her general pediatrician for possible psoriasis. The patient presented complaining of a widespread, pruritic rash for 1 year. Aside from these symptoms, she reported feeling well. Prior to presenting at the dermatology clinic, she had received treatment with oral cephalexin and oral cetirizine for working diagnoses of folliculitis and atopic dermatitis, respectively. The patient's mother reported that these treatments did not improve her condition. Prior use of CeraVe and topical hydrocortisone treatments also failed to control the patient's symptoms.

On physical exam, hundreds of coalescing, monomorphic papules were found on the face, trunk, extremities, and intertriginous areas *(Figure 1a)*. Lesions varied between scaly and shiny, and some linear papules due to koebnerization were found on the right arm *(Figure 1c)*. Based upon clinical presentation, the patient was diagnosed with lichen nitidus and atopic dermatitis. Further inquiry revealed that the pruritus was limited to areas of the body appearing to be affected by atopic dermatitis (specifically, the nape of the neck and arms), and areas with a higher concentration of lesions appearing to be lichen nitidus (specifically, the face and trunk) were minimally pruritic. Topical hydrocortisone 2.5% ointment and topical triamcinolone 0.1% ointment twice daily were prescribed as needed when her symptoms worsened.

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The patient's mother gave permission for publication of this case.
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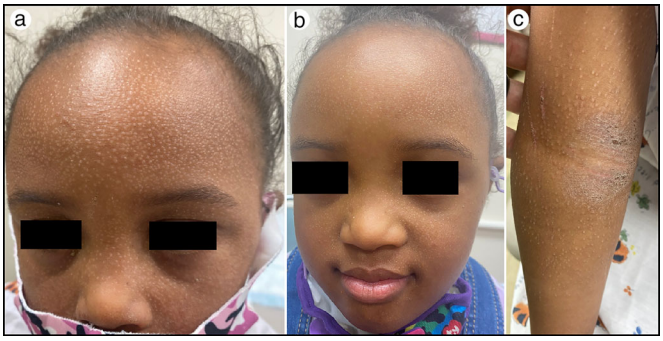

Figure 1. **(a)** Patient's facial lichen nitidus lesions. Note the widespread distribution. **(b)** Patient's facial lesions following 3 months of treatment. **(c)** Patient's right arm. Note the Koebner phenomenon and eczema at antecubital fossa.

After 3 months of hydrocortisone and triamcinolone treatment several times per week, many of the patient's papular lesions improved, reducing to postinflammatory hypopigmentation with some papules remaining *(Figure 1b)*. The patient was counseled to continue using the prescribed treatment as needed. Because the patient and her mother are pleased with her progress, the patient's condition will continue to be monitored for worsening symptoms. If the patient's lichen nitidus is not well controlled in the future, topical calcineurin inhibitors will be considered because they provide a relatively more sustainable treatment option.

DISCUSSION

The extensive number and distribution of papules on this patient's face and elsewhere on the body is unusual for lichen nitidus, though the morphology of discrete, shiny, flat-topped, grouped, and koebnerized papules allowed for a clinical diagnosis. Because of the patient's young age, a biopsy was not performed. Classically, histopathology reveals a "ball in clutch" or "ball and claw" of epidermal rete around a lymphohistiocytic infiltrate (sometimes with multinucleated giant cells). There is parakeratosis overlying epidermal atrophy and focal basal liquefaction degeneration.[10,11]

Several case reports demonstrate an association of lichen nitidus with Down syndrome.[3–9] Based on a search of PubMed, Scopus, and Embase, of the cases of lichen nitidus reported in patients with Down syndrome, most (seven of eight, including this patient) *(Table 1)* describe a generalized distribution of lesions. By documenting another case of generalized lichen nitidus in a patient with Down syndrome, we provide additional evidence suggesting a relationship between these conditions. *Table 1* also characterizes the age of initial presentation and the gender of previously described cases. Pathophysiologic determinants leading to a more generalized presentation of lichen nitidus in patients with Down syndrome will require further exploration.

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Table 1. Reported patients with lichen nitidus and Down syndrome

Cases	Age (years) at presentation	Sex	Distribution
Patrizi et al[3]	9/12	M	Generalized, sparing scalp, palms, soles
Prigent et al[4]	2	F	Initially, posterior neck and right wrist; slow progression to trunk, neck, buttocks, and right wrist
Laxmisha and Thappa[5]	2	F	Abdomen initially, progressing to involvement of trunk, face, genitalia, upper and lower limbs
Henry and Metry[6]	3	M	Generalized with perioral and perinasal accentuation
Botelho et al[7]	4	F	Face, trunk, upper and lower limbs, genitalia
Guliani et al[8]	4	M	Wrists initially, progressing to generalized
Agarwal et al[9]	4	F	Extensor surface of legs and thighs
Present case	6	F	Face, trunk, upper and lower limbs, intertriginous areas

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Avocations

"South Africa—Family Going Home" by Jay Hoppenstein, MD. Dr. Hoppenstein (e-mail: navigato@aol.com) is a surgeon who was on the medical staff at Baylor University Medical Center at Dallas from 1971 until his retirement in 2005. He also served as chairman of the Department of Surgery at Presbyterian Hospital of Dallas.